

SURGERY FOR CONGENITAL HEART DISEASE

WHAT FACTORS AFFECT VENTRICULAR PERFORMANCE AFTER A FONTAN-TYPE OPERATION?

Postoperative conditions after a Fontan-type operation, particularly as they affect results in the early term, are thought to depend on factors such as the state of pulmonary circulation and ventricular function. In this study, we attempted to determine the factors that influence ventricular characteristics in the middle term after Fontan-type procedures. Catheterization was performed at a mean of 15 months after operation in 57 patients with univentricular atrioventricular connection who underwent the operation between 1.0 and 22.6 years of age. End-diastolic volume, end-systolic volume, ejection fraction, and end-diastolic pressure of the systemic ventricle were analyzed together with an estimation of the systemic flow index. These parameters were influenced significantly by the presence of atrioventricular valve insufficiency. The morphologically left ventricle showed a better ejection fraction than did the morphologically right ventricle, whereas the systemic flow index was greater in patients undergoing total cavopulmonary connection than in those receiving an atriopulmonary connection. Young age was significantly associated with a better postoperative contractility, whereas the potential for impaired ventricular compliance was suggested in several patients undergoing operation after 4 years of age. On the basis of our results, we conclude that total cavopulmonary connection performed at a young age should be the surgical procedure of choice and that atrioventricular insufficiency must be treated properly at, and even after, the initial definitive repair. (*J THORAC CARDIOVASC SURG* 1995;110:405-15)

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Subsequent to the increasing number of successful experiences with Fontan-type operations, several factors have been shown to influence operative

results.¹⁻³ On this basis, alternative surgical strategies have been devised, either to improve results still further or to extend the indications of this functionally definitive repair procedure.⁴⁻¹¹ Other recent investigations have shown that factors such as the occurrence of atrial arrhythmias and the response of cardiopulmonary function to exercise have a major influence in the long term.¹²⁻²⁰ In this light, it seems reasonable to suggest that either ventricular function or the state of the pulmonary circulation could have major influences not only on the outcome immediately after a Fontan-type operation but also on the functional status in the longer term. With these possibilities in mind, we have attempted to identify factors which might influence postoperative

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Table I. Morphologic conditions (No. of patients)

	Absent AV connection		Double inlet	
	Absent right	Absent left	To morph. RV	To morph. LV
Usual atrial arrangement	24	6*	4† 0‡	3† 0‡
Right isomerism		2	2† 12‡	0† 3‡
Left isomerism			0† 1‡	
Dominant morph. LV	24	1		6
Dominant morph. RV		7	19	

AV, Atrioventricular; *morph.*, morphologically; RV, right ventricle; LV, left ventricle.

*Including one with discordant AV connection with an imperforate left-sided AV valve.

†Two AV valves.

‡Common AV valve.

ventricular performance in the middle term, using data derived from postoperative catheterizations.

Patients and methods

Since 1980, 124 patients (with classic tricuspid atresia in 39 and other anatomic arrangements in 85) underwent Fontan-type operations, excluding the so-called fenestrated Fontan procedure,²¹ at the National Cardiovascular Center in Osaka, Japan. Of the 104 operative survivors (35 and 69 from the two groups, respectively), 79 patients have thus far undergone postoperative catheterization along with ventriculography (24 and 55 from each group, respectively). Among these, we selected for study those patients unified by the morphologic condition of a univentricular atrioventricular connection, thus excluding 22 patients with biventricular atrioventricular connections (those with pulmonary atresia with intact ventricular septum and those with double outlet right ventricle with common atrioventricular valve, for instance). Fifty-seven patients were left within our study group.

Usual atrial arrangement was seen in 37 patients, isomerism of the right atrial appendages was seen in 19, and left isomerism was seen in one. Of the 37 patients with usual atrial arrangement, the right atrioventricular connection was absent in 24 (producing the classic pattern of tricuspid atresia) and the left connection was absent in six. In the other seven patients, there was double inlet to the morphologically right (four patients) or to the morphologically left ventricle (three patients), all with the atrioventricular junctions guarded by two separate atrioventricular valves. All patients with isomeric atrial appendages, except two whose right atrioventricular connection was absent, had double inlet atrioventricular connections to the morphologically right ventricle in 15 patients and to the morphologically left ventricle in three patients. The junctions were guarded by two separate atrioventricular valves in two patients with double inlet right ventricle but by a common atrioventricular valve in the other 16. Overall, 31 patients showed a dominant morphologically left ventricle with an incomplete and rudimentary right ventricle, and 26 patients showed a dominant morphologically right ventricle with an incomplete and rudimentary left ventricle (Table I).

Seven patients had previously undergone banding of the

pulmonary trunk with (three patients) or without (four patients) reconstruction of the aortic arch for either coarctation or interruption of the aorta. The interval between the palliative procedure and the Fontan-type operation ranged from 1.7 to 9.5 years (mean 3.5 ± 2.7 years). Previous anastomoses between the superior caval vein and the pulmonary arteries had been achieved in another seven patients by means of a conventional Glenn procedure (five patients) or bidirectional cavopulmonary anastomosis (two patients). These palliative operations had been performed 2.3 ± 0.9 years before the total right heart bypass operation. Of the other 43 patients, a systemic-to-pulmonary arterial shunt had been constructed in 19 patients, repair of totally anomalous pulmonary venous connection carried out in two, atrial septectomy done in one, and the Norwood procedure done in one.

The definitive surgical procedure was done in patients from 1.0 to 22.6 years of age (mean 6.3 ± 4.2 years) (Fig. 1) by means of either an atriopulmonary connection²² in 21 patients, a modified atriopulmonary connection along with the anastomosis of the superior caval vein to the pulmonary arteries in four, or some modification of the total cavopulmonary connection in 32²³ (Table II). Each method was achieved with the use of crystalloid cardioplegic solution for cardiac arrest. We established total cavopulmonary connection by intraatrial rerouting either with a polytetrafluoroethylene tube in 17 patients or with a heterologous pericardial baffle in 15 patients, anticipating the growth of their own atrial walls to provide for an effective pathway in the future. These methods provide venous drainage from the heart itself to a low-pressure atrial chamber which is connected to the systemic ventricle, and also respect the site of the sinus node²⁴ (Fig. 2). The width of the native tissue included within the pathway was designed to be no more than one third of the circumference at any cross section of the created route. In this way, it was intended to minimize any hemodynamic effects of atrial contraction which may have produced reflux back to the caval veins.²⁵

At the time of the Fontan-type procedure, extensive plasty of the anulus or the leaflets was performed in eight patients with a common atrioventricular valve, whereas the morphologically tricuspid valvular orifice was closed in

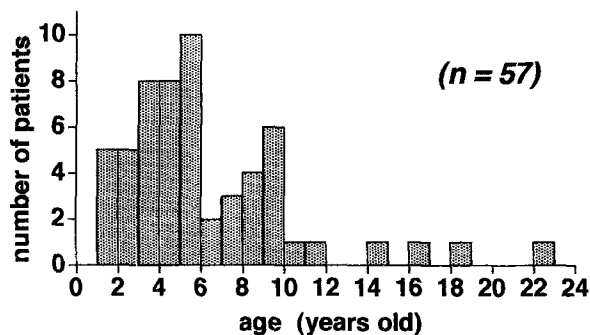


Fig. 1. Age at the time of definitive operation.

four patients with two separate valves (Table II). These additional surgical procedures were considered necessary because moderate to severe regurgitation across the atrioventricular valve had been recognized before operation. Recurrent insufficiency was seen in two patients, one with a common valve and the other with a solitary morphologically tricuspid valve. In the other nine patients, the treatments were successful. In only one patient was moderate regurgitation across the common valve left untreated. Residual moderate regurgitation neither progressed nor regressed. Of the 45 patients in whom valvular repair was deemed unnecessary because of the preoperative finding of no more than slight insufficiency, three patients with absence of either the right or left atrioventricular connection had progressive regurgitation of the atrioventricular valve after Fontan-type procedures. The regurgitant atrioventricular valve was the morphologically tricuspid valve in two patients and the morphologically mitral valve in the third patient. Anastomosis of the proximal pulmonary trunk to the ascending aorta was performed at the time of the Fontan-type operation to avoid a possible subaortic stenosis in four of seven patients who had previously had banding of the pulmonary trunk.^{26, 27} Simultaneous repair of totally anomalous pulmonary venous connection was performed in five patients with isomerism of the right atrial appendages.

Postoperative catheterization was done 15.0 ± 8.5 months (range 2 to 52 months) after the operation. Among data derived from the examination, we selected three parameters by ventriculography, namely, end-diastolic volume, end-systolic volume, and ejection fraction of the dominant ventricle supporting the systemic circulation, in addition to a measured value of end-diastolic pressure of the dominant ventricle and an estimated value of the quantity of systemic flow.

Volumetry was done by means of ventriculography (direct injection into the ventricle only) with biplane projections and calculated by means of an integration technique (Simpson's method or area-length method) along with proper revisions for either the morphologically right or left ventricle.²⁸ For standardization, the calculated end-diastolic and end-systolic volumes were divided by the anticipated value of end-diastolic volume in the normal heart. Values for dominant morphologically left ventricles were compared with those of the normal left

Table II. Options performed at the time of definitive repair (No. of patients)

	Usual arrangement	Isomerism
Method of the Fontan-type procedure		
APC	20	1
APC with bidirectional cavopulmonary anastomosis	4	0
TCPC using a tube graft	2	15
TCPC using a baffle	11	4
Simultaneous surgical procedures		
Plasty of the common atrioventricular valve	0	8
Closure of the morphologically tricuspid valve	4	0
Additional aortopulmonary anastomosis	4	0
Repair of totally anomalous pulmonary venous connection	0	5

APC, Atriopulmonary connection; TCPC, total cavopulmonary connection.

ventricles, and the dominant right ventricle was compared with the normal right ventricle. All values were given in percentages. Each normal value of end-diastolic volume was calculated from body surface area.²⁸ End-diastolic pressure (in millimeters of mercury) was represented by the average of 10 consecutive recorded values. The amount of systemic blood perfusion was determined by means of the Fick oxymetric method, from oxygen consumption and oxygen content within pulmonary and systemic arterial blood, and its index (per body surface area, in liters per minute per square meter) was used for analysis. In patients with an anastomosis between the superior caval vein and the pulmonary arteries, including those treated with total cavopulmonary connection, a mean value of the oxygen content in the left and right pulmonary arteries was used as an estimate of the real value of mixed venous oxygen content. Additionally, we divided the ventricular end-diastolic pressure by the index of systemic flow when the volume load altered by atrioventricular regurgitation was trivial, expecting that the derived value might serve as an estimation of ventricular diastolic compliance.²⁹

Statistical analysis was achieved with the use of the F-test, unpaired *t*-test, and regression analysis for data derived from postoperative catheterization, as well as logistic regression for multivariate analysis of influence of clinical factors.

Results

Values of end-diastolic volume, end-systolic volume, and end-diastolic pressure were significantly greater in patients with moderate or severe regurgitation than in those without and were accompanied with smaller indexes of systemic flow. The ejection fractions of the ventricle were reduced in three of six patients with insufficiency—these values falling out-

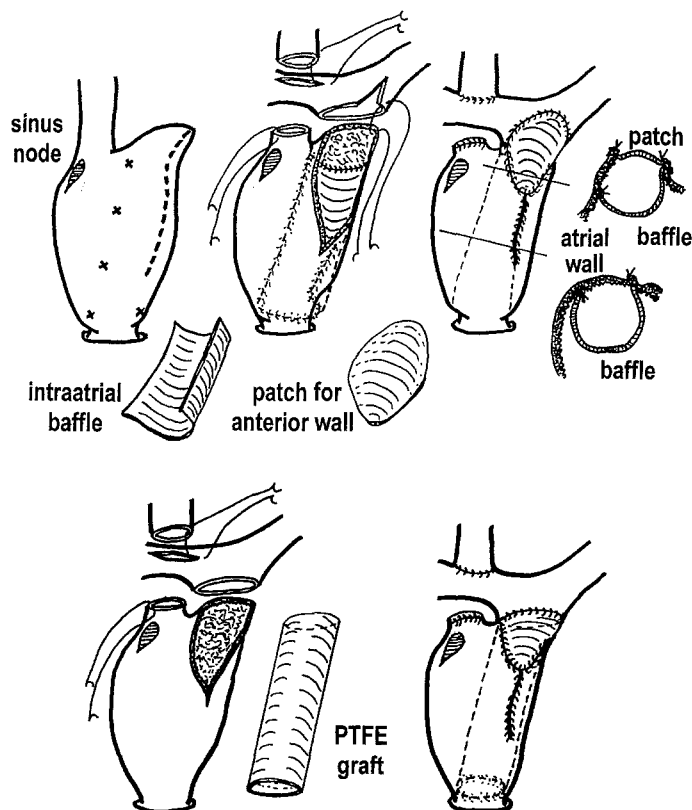


Fig. 2. Operative method used for achieving total cavopulmonary connection. The sinus node and cardiac venous drainages were placed within the low-pressure atrial chamber connected to the systemic ventricle. Part of this figure was redrawn from the illustrations from Yagihara and associates.²⁴ PTFE, Polytetrafluoroethylene.

side the range of one standard deviation around the mean for the patients without regurgitation. Reparative procedures performed on the atrioventricular valves at the time of the Fontan-type procedure themselves, if effective, produced no difference in these characteristics (Fig. 3).

Excluding these six patients with significant regurgitation, a statistically significant influence of the morphologic condition of the dominant ventricle was detected only for ejection fraction (Fig. 4). Patients undergoing total cavopulmonary connection had a greater index of systemic flow (2.87 ± 0.37 L/min per square meter) than did those receiving an atriopulmonary connection (2.29 ± 0.39 L/min per square meter) (Fig. 5). Analysis of other ventricular characteristics showed no differences between the two groups. Furthermore, ventricular characteristics were not influenced by either the Glenn procedure or by banding of the pulmonary trunk before the definitive repair.

As for the effect of age at operation, an inverse correlation was found between age at definitive

operation and ejection fraction ($r = -0.57$, $p < 0.001$) (Fig. 6). Correlations were not apparent between age at operation and other parameters.

Interrelation of clinical factors were shown within morphologic conditions or between morphologic conditions and surgical options performed but were not identified between or within other factors (Table III). Logistic regression showed similar outcomes as seen with the univariate analyses (Table IV). Stepwise procedure for multivariate regression showed that the influence of age at operation on ejection fraction was independent of those of other clinical factors.

Discussion

Surgical mortality after a Fontan-type procedure continues to improve; now, either the long-term postoperative results or subsequent functional status is regarded as the crucial matter of concern.¹³⁻²⁰ Because the Fontan circulation lacks any driving pressure for pulmonary perfusion produced by ventricular contraction, optimal states of the pulmonary circulation and

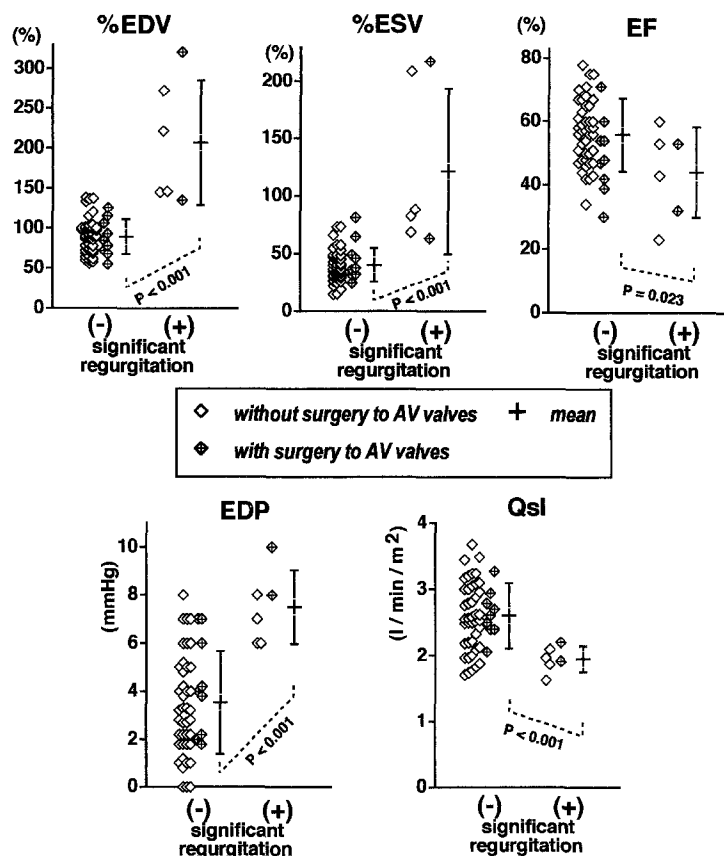


Fig. 3. Influence of significant atrioventricular valve regurgitation. %EDV, %ESV, End-diastolic and end-systolic volumes (respectively) of the systemic ventricle compared with the anticipated normal value calculated from patient's body surface area; EF, ejection fraction; EDP, end-diastolic pressure of the systemic ventricle; QsI, quantity of effective systemic perfusion divided by body surface area; AV, atrioventricular; -, slight or no regurgitation; +, moderate or severe regurgitation.

ventricular function can reasonably be considered as fundamental factors for an overall optimal circulation. In this study, we focused on ventricular characteristics derived from catheterization to assess the physiologic performance of the ventricular mass after total right heart bypass operations. Parameters derived from other examinations, such as echocardiography or ventricular pressure-volume studies, should be equally useful in determining precisely any impaired diastolic ventricular function.³⁰

A limitation of our investigation lies in the selection of the patients. The survivors of surgical procedures might have had fewer risk factors for the achievement of the definitive surgical procedure. Accordingly, some of the factors which portend a poor prognosis might have been excluded simply by our selection of survivors. In contrast, there is probably no bias in the population in whom postoperative catheterization was performed. It is our policy that postoperative catheterization is done

routinely from 1 to 2 years after a Fontan-type procedure. All survivors from our center with univentricular atrioventricular connection undergoing operation before February 1993, therefore, were included in this study, excluding three whose ventriculograms were unavailable for analysis.

Patients with the univentricular atrioventricular connection were selected to minimize the equivocal estimation of the ventricular volumes in those with two balanced ventricles. When a heart possesses biventricular atrioventricular connections and balanced morphologically right and left ventricles, the measured ventricular volume cannot be standardized by simple comparison with either the right or the left ventricle of the normal heart. We presume the presence of the dominant ventricle with an incomplete and rudimentary ventricle should considerably diminish errors in measurement. Yet, it remains controversial to compare values of the right and left ventricular volume divided by each normal value. It is

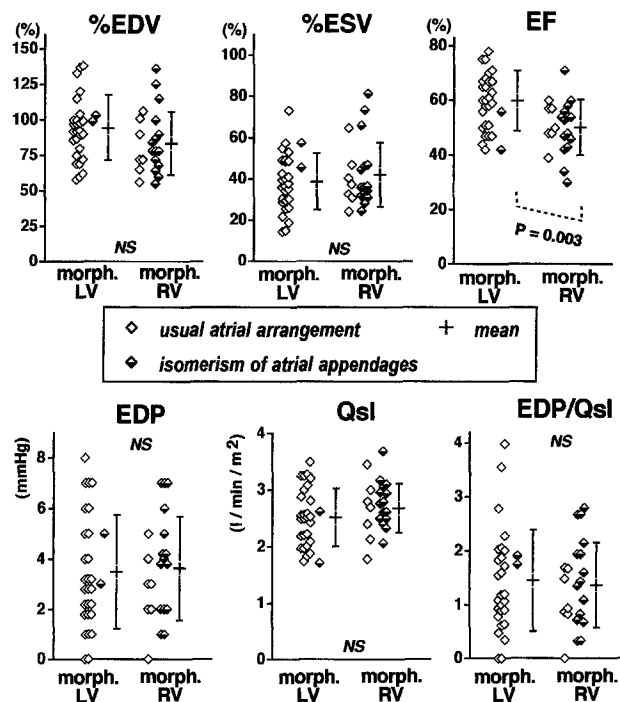


Fig. 4. Difference between the morphologically left and right ventricles. *morph.*, Morphologically; *LV*, left ventricle; *RV*, right ventricle. For other abbreviations, see Fig. 3.

likely acceptable, nonetheless, to compare these two different subsets of values on the same stage.

Another problem may exist in the estimation of the quantity of systemic flow. The anastomosis between the superior caval vein and the pulmonary arteries makes it impossible to obtain the true value of oxygen content in mixed venous blood. It is an approximation of true systemic flow which we have substituted by using the mean of values in the right and left pulmonary arteries for estimation of the mixed venous content of oxygen. This alternative method is based on the unreal assumption that the pulmonary flow is equal on each side. It remains, nonetheless, one of the simple ways available for evaluating the systemic flow by oxymetry in this setting. Division of end-diastolic pressure by the index of systemic flow is similarly imprecise. This value was held to have the probability of adversely representing the diastolic ventricular compliance.²⁹ The concept itself seems theoretically sound. As yet, we are unable to determine whether its application to our data is appropriate.

The final limitation of our study is the circumstance of the catheterization itself. The examination was performed with the patient under sedation, conditions obviously different from the resting state

or exercise. As has been well established, after a Fontan-type operation patients show impaired cardiopulmonary responses to exercise, such that the parameters measured at rest do not necessarily reflect functional status.¹⁵⁻²⁰ All our values, therefore, may offer only information about "static" physiology and cannot predict completely the dynamic functional status of the circulation.

These limitations in material and methods in turn limit the making of decisive comments on our results. Nonetheless, the deleterious effect of atrioventricular insufficiency was unequivocal. Atrioventricular regurgitation is known to be among the risk factors negating the achievement of a successful Fontan circulation² and is also among the causes of late death.¹² Our physiologic data support similar conclusions in the middle term. Moderate to severe insufficiency of the atrioventricular valve, either recurrent or new, was associated with poorer ventricular performance, as well as with lower systemic perfusion. Such disastrous regurgitation is likely to occur more frequently across solitary tricuspid or common atrioventricular valves but is unlikely to be related to ventricular structure. In all patients but one, the regurgitation was detected to be less than mild during some period of their postoperative

courses. We suggest, therefore, that progressive regurgitation should be diagnosed and treated properly before ventricular dysfunction becomes irreversible, even after the definitive surgical procedure.

Ventricular morphologic condition could be one of the most crucial factors likely to affect physiologic parameters. The unsuitability of the morphologically right ventricle for the systemic circulation has been reported both in congenitally corrected transposition and in complete transposition (concordant atrioventricular and discordant ventriculoarterial connections) after the atrial switch procedure. It depends presumably on differences in anatomic structure between the ventricles. A similar deterioration seems possible in patients with a dominant morphologically right ventricle after a Fontan-type operation.^{31, 32} No statistical differences, apart from ejection fraction, however, were found between the groups of patients with dominant morphologically right and left ventricles. This finding suggests that ventricular characteristics are potentially subject to the pulmonary circulation. Even the difference seen in ejection fraction cannot be used to determine whether it has clinical significance in contraction. The normal morphologically right ventricle is known to show smaller values for ejection fraction than the comparable normal morphologically left ventricle when estimated with the same methods as applied in this study ($60\% \pm 6\%$ versus $65\% \pm 5\%$).²⁸ We must, therefore, limit our comments to state that deterioration in function of the dominant right ventricle was not clearly detected from our results obtained in the middle term. Further investigations in both the longer term and during exercise are needed to clarify the behavior of the morphologically right ventricle when supporting the systemic circulation. Judging from the lack of evidence relating ventricular morphologic condition to age at operation, we believe that age itself did not contribute to the statistical difference between two groups of patients with differing ventricular morphologic condition.

With regard to variations in surgical options, loss of hydrodynamic energy during passage of blood through the pathway from the caval veins to the pulmonary arteries,²³ a reduced coronary arterial flow caused by a raised pressure within the coronary sinus,³³ and higher incidence of postoperative atrial arrhythmias probably caused by incorporation of the sinus node in the high-pressure atrium¹⁴ have all been reported as factors which potentially influence ventricular performance or affect the Fontan circu-

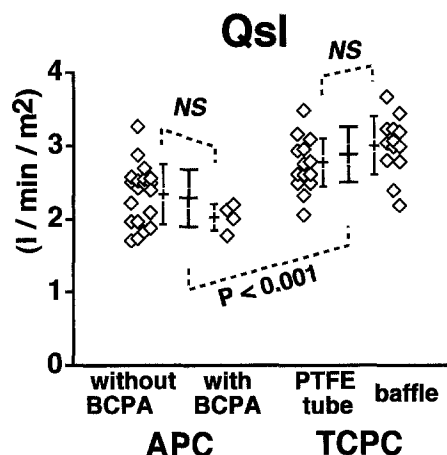


Fig. 5. Influence of operative methods. Quantity index of systemic perfusion was significantly greater in the setting after total cavopulmonary connection than after atriopulmonary connection. *QsI*, Quantity of effective systemic perfusion divided by body surface area; *PTFE*, polytetrafluoroethylene; *BCPA*, bidirectional cavopulmonary anastomosis; *APC*, atriopulmonary connection; *TPC*, total cavopulmonary connection; *NS*, not significant.

lation itself. Our technique for total cavopulmonary connection has been devised to minimize the energy loss and to place both the cardiac venous drainage and the sinus node within the low-pressured atrial chamber connected to the systemic ventricle²⁴ (Fig. 2). When an intraatrial baffle was used, the anterior wall of the right atrium was used instead of the lateral part and the atrial septum, as are used in the lateral tunnel method.²³ Although further investigations are obviously necessary regarding the precise contribution of these modifications, the preferable systemic flow index presently observed in patients undergoing total cavopulmonary connection may well reflect the efficiency anticipated from these minor modifications.

Finally, the influence of age at operation on ventricular characteristics can be assumed on the basis of the histopathologic change induced by cyanosis.^{34, 35} Impaired ventricular function is seen more often in patients after a later biventricular repair in tetralogy of Fallot³⁶ and in other malformations.³⁷ In the Fontan circulation, it is still unclear whether ejection fraction is directly correlated to ventricular contractility because preload to the ventricle is reduced in the unusual pulmonary circulation lacking the impact of a pulmonary ventricle. In addition, age at catheterization may also affect the ejection fraction. Some authors mentioned the

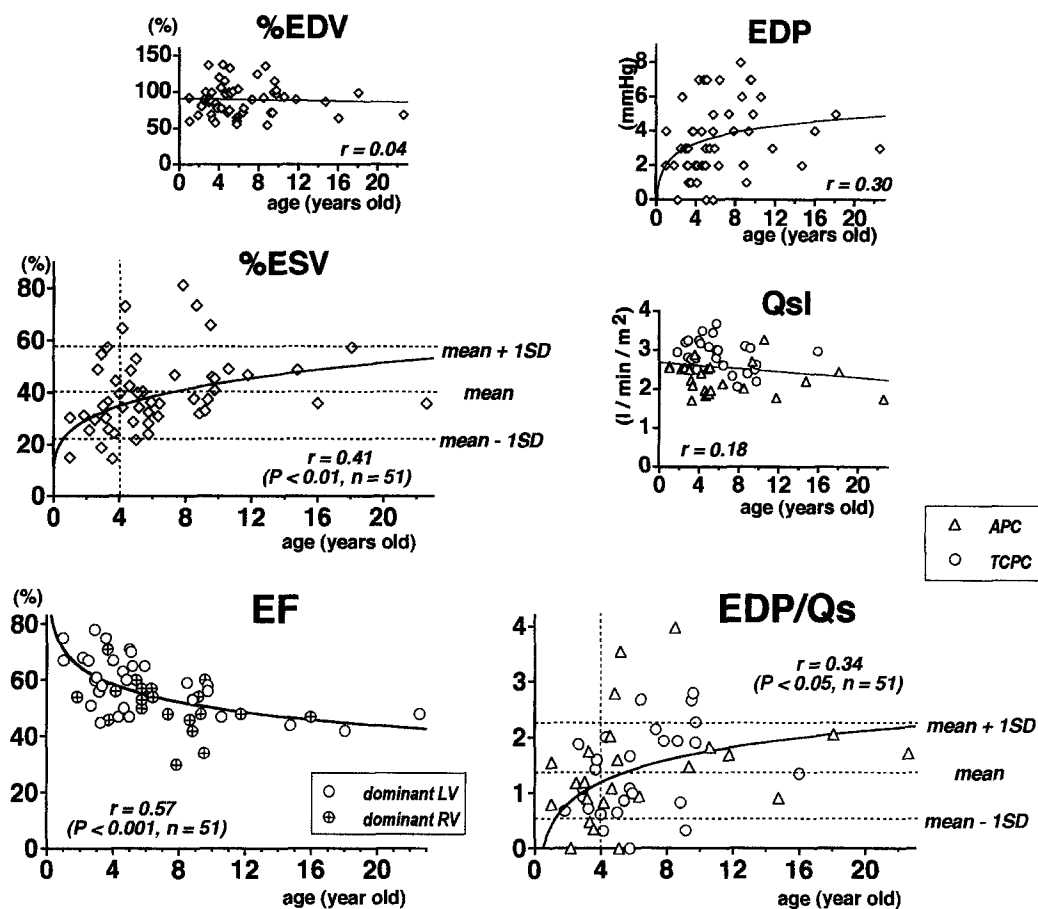


Fig. 6. Influence of age at definitive procedure. APC, Atriopulmonary connection; TCPC, total cavopulmonary connection; LV, left ventricle; RV, right ventricle. For other abbreviations see Fig. 3.

Table III. Interrelation between clinical factors (correlation coefficients)

	Morphologic condition			Postoperative regurgitation of AV valve	Surgical option			Age at definitive operation
	Atrial arrangement	Dominant ventricle	AV valve		Method for Fontan type procedure	Option for AV valve	Previous operation	
Morphologic condition								
Atrial arrangement	—	0.58*	0.84*	0.11	0.67*	0.48*	0.03	0.10
Dominant ventricle	0.58*	—	0.74*	0.14	0.59*	0.46*	0.13	0.06
AV valve	0.84*	0.74*	—	0.20	0.68*	0.49*	0.07	0.12
Postoperative regurgitation of AV valve	0.11	0.14	0.20	—	0.23	0.16	0.17	0.10
Surgical option								
Method for Fontan type procedure	0.67*	0.59*	0.68*	0.23	—	0.31	0.21	0.03
Previous operation	0.48*	0.46*	0.49*	0.16	0.31	—	0.25	0.00
Option for AV valve	0.03	0.13	0.07	0.17	0.21	0.25	—	0.00
Age at definitive operation	0.10	0.06	0.12	0.10	0.03	0.00	0.00	—

There were relationships between and within the morphologic conditions and options used, but postoperative recurrence of atrioventricular valve regurgitation, previous palliative operations, and age at definitive procedure were independent factors. AV, Atrioventricular.

* $p < 0.001$ ($n = 57$).

Table IV. Influence of clinical factors on parameters derived from catheterization (*p* values for logistic regression)

	%EDV	%ESV	EF	EDP	QsI	EDP/QsI
Morphologic condition						
Atrial arrangement	NS	NS	0.020	NS	NS	NS
Dominant ventricle	NS	NS	0.009	NS	NS	NS
AV valve	NS	NS	0.006	NS	NS	NS
Postoperative regurgitation of AV valve	<0.001	<0.001	0.023	<0.001	0.002	<0.001
Surgical option						
Method for Fontan type procedure	NS	NS	0.023	0.011	0.001	0.033
Previous operation	NS	NS	NS	NS	NS	NS
Option for AV valve	NS	NS	NS	NS	NS	NS
Age at definitive operation	NS	NS	0.014	NS	NS	NS

Logistic regression analysis showed the outcomes similar to those shown by univariate analyses. %EDV, %ESV, End-diastolic and end-systolic volumes (respectively) of the systemic ventricle compared with the anticipated normal value calculated from patient's body surface area; EF, ejection fraction; EDP, end-diastolic pressure of the systemic ventricle; QsI, quantity of effective systemic perfusion divided by body surface area; NS, not significant; AV, atrioventricular.

possibility of correlation between age at examination and ejection fraction,³⁶ but others showed negative findings.²⁸ In our series, earlier definitive repair corresponds to younger age at postoperative catheterization. It is not easy to define the exact advantage of a good ejection fraction in the Fontan circulation, but a better fraction can never be considered to worsen the efficiency of the circulation. Our data from both end-systolic volume and the division of end-diastolic pressure by the quantity of systemic flow show a tendency toward impaired ventricular function as a consequence of later definitive repair. Values of end-systolic volume more than the mean plus one standard deviation were recognized in patients who underwent definitive surgical procedures when older than 4 years of age. Similarly, the division of end-diastolic pressure by the quantity of systemic flow may indicate the stiffness of the ventricular mass in older patients, values larger than a mean plus one standard deviation being plotted again in those older than 4 years of age. These effects of age at definitive repair could be eliminated only by proceeding to earlier definitive surgical procedures.

Nonetheless, two major hazards militate against an earlier Fontan-type procedure. The first hazard lies in age itself as a risk factor. A young age has long been held to be associated with less successful operative outcome, both in the short and the long terms.^{1-3, 8-10, 12} This factor has been neutralized to some degree by recent experiences. In one report, for example, the authors have gone so far as to recommend an earlier Fontan-type procedure for better results.¹¹ Still, the effect of extending the age-related indication is limited because of the

special situation of this postoperative circulation. A Fontan-type procedure during neonatal life or early infancy, for example, is far from a reasonable surgical choice, at least at the present time. This scenario is markedly different from early biventricular repair in tetralogy of Fallot or other malformations. The findings of our study, therefore, should not be interpreted to suggest that earlier definitive repair provides a better prognosis. Rather, we would argue that it is not always justifiable to postpone definitive surgical procedures when the only reason for delay is that the patients should be 2 or 3 years of age. In other words, younger age can be advantageous with regard to overall surgical outcome if the other conditions are suitable for the Fontan circulation and conducive to low operative mortality.

The other hazard contraindicating earlier operation is the probability of obstruction within the pathway constructed from the caval veins to the pulmonary arteries. Our preference in surgical options is the total cavopulmonary connection. The use of a prosthetic graft in such a procedure could result in obstruction either by its lack of potential for growth or because of the formation of thrombus within it. The alternative method, using a baffle, can be one of the methods of choice in which the growth potential of the pathway is respected. Obviously, another long-term investigation is required to settle this matter. The other techniques for establishing the Fontan circulation are also exposed to the risk of future obstructions, and younger age, as well as smaller body size, may accelerate the development of such stenosis. We conclude that optimal age for the Fontan-type operation must be determined for each patient with respect to both advantages and disadvantages of an earlier operation.

In summary, although the findings of our study impinge on only a small part of the long-term results of the Fontan-type procedure, they support our basic policy that the total cavopulmonary connection is a reasonable surgical option and that delayed operation does not necessarily mean better results in survivors. Simultaneous treatment of a regurgitant atrioventricular valve has been shown to be feasible at the time of definitive surgical procedures and has had no disadvantageous effects on postoperative ventricular characteristics. If moderate regurgitation of the atrioventricular valve occurs after the operation, even if only moderate, it can have a great influence on subsequent progress. In such circumstances, additional valvular operations would seem to be indicated.

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Discussion

Dr. Davis C. Drinkwater, Jr. (*Los Angeles, Calif.*). I enjoyed your talk very much. We have been using postoperative captopril therapy for every patient with a Fontan procedure. We believe that this therapy makes a difference over the long term, although we do not have any definitive data at this point. Do you have any particular postoperative medical management that you have developed for these patients?

Dr. Uemura. You mean medication?

Dr. Drinkwater. Yes, such as captopril—afterload reducing to prevent atrioventricular valve regurgitation and future dilatation, particularly in the right ventricular morphology?

Dr. Uemura. Usually we use medication such as diuretics or digoxin in addition to warfarin as an anticoagulant, but not dilators. The patients with postoperative regurgitation of the atrioventricular valve tend to receive larger doses of furosemide or other diuretics.

Dr. Richard A. Jonas (*Boston, Mass.*). I do not think any of us would disagree with the proposition that the earlier you can eliminate the volume load on a single ventricle, which is inherent in having a pulmonary artery band or an arterial shunt-dependent-type circulation, the better, but I am not sure that necessarily tells us that a completed Fontan operation is preferable to other methods of eliminating that volume load, such as the bidirectional Glenn shunt. The bidirectional Glenn shunt eliminates pulmonary recirculation and does not have the disadvantages of increasing hepatic venous pressure, mesenteric venous pressure, or right atrial pressure. In view of the fact that the late complications of the Fontan operation are cirrhosis, protein-losing enteropathy, and arrhythmias, perhaps we should be considering several years of palliation with a bidirectional shunt followed subsequently by a Fontan operation. Whether the addition of a fenestration will be an advantage with regard to reducing those long-term deleterious effects is also unclear.

Do you have any information or any sense as to the comparative palliation achieved with the bidirectional Glenn versus fenestrated Fontan versus completed Fontan?

Dr. Uemura. Our policy is to achieve complete relief of cyanosis. Accordingly, we usually choose a complete Fontan type of operation, if possible, instead of a bidirectional Glenn procedure. After a conventional bidirectional Glenn shunt, the pulmonary arterial sizes may regress because the total amount of pulmonary flow decreases after the procedure. Therefore, in our hospital, a bidirectional Glenn procedure is established together with banding of the pulmonary trunk or with leaving the pulmonary valve open. In other words, we maintain the blood flow from the ventricle to the pulmonary arteries so that volume reduction by the operation is not as complete as a total right heart bypass operation or a bidirectional Glenn shunt. Our preference is, therefore, to use a Fontan-type operation, aiming to offload the ventricle adequately. Because of these strategies, we have no information regarding other procedures such as a fenestrated Fontan or a conventional bidirectional Glenn shunt.